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Case Report

Metastatic gastrointestinal stromal tumours- a rarest of rare presentation: a case report

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ABSTRACT

Gastrointestinal stromal tumours (GISTs) is the most common mesenchymal tumour of the gastrointestinal tract (GIT). GISTs can occur anywhere along the GIT, more so commonly in the stomach and small intestine. They can manifest as an emergency such as obstruction, perforation of a hollow viscous or as a haemorrhage. The metastatic stage was usually detected after the histopathologic report. We report on a 29-year female para 1 live 1 (P1L1) admitted at the institute of obstetrics and gynaecology (IOG) Egmore, Chennai, as a case of right torsion ovary. The patient was taken for an emergency laparotomy. Ovaries are found to be normal and so the surgical team was called in. Per-operative diagnosis of ileo-ileal intussusception was made, followed by resection and anastomosis. Biopsy report of the specimen showed a GIST, arising from the small intestine causing the intussusception. Further follow-up of the patient with CECT abdomen showed hepatic metastasis in segments 4a and 7. GISTs tumours data on worldwide frequency is limited, especially in the 3rd world countries. Adding to the limited data GISTs present themselves in protean ways. It is then necessary to understand not only the presentation but also the complications. Multimodality approach involving early screening, dissemination of knowledge regarding various types of presentation and the tools to manage such complications, early involvement of medical gastroenterology and medical oncology along with patient education will go a long way in the management of these difficult tumours.

Keywords: Gastrointestinal stromal tumours, Intessusception, Mesenchymal tumours

INTRODUCTION

GISTs arise from special cells in the wall of the GIT named interstitial cells of Cajal (ICCs). ICCs are the pacemakers of the GIT, as they signal the muscles in the GI tracts to contract, to move food and liquid forward. These are mainly bipolar cells or spindle-shaped cells associated with the long axis of the surrounding smooth muscle cells. These cells do not form their network.[1] Cajal (1852-1934) is considered to be one of the founders of the field of neuroscience. In 1911, he described interstitial neurons in the gut, noting that they were primitive accessory components that perhaps modify smooth muscle contraction, themselves subject to regulation from principle neurons. The accuracy of his description of their appearance and activities has led to these cells now being called the ICC.[2]

Many GISTs are discovered incidentally during endoscopic or surgical procedures. Other GISTs are detected on radiologic studies performed to investigate protean manifestations of GIT disease or procedures performed to treat an emergent condition such as hemorrhage or obstruction. Based on the study in western Sweden the median tumor size for incidental findings was measured to be 2.7 cm and that of the tumors found based

on symptoms was 8.9 cm.[3] These tumors have been reported to range in size from smaller than 1 cm to as large as 40 cm in diameter.[4]

Most of the GISTs originate in the stomach ranging from 50-70% and the small intestine ranks second most common location comprising 20-30% of GISTs. Less frequent sites include the colon, rectum and oesophagus. Extra GI sites include pancreas, omentum, and mesentery, but very rarely reported.[5] Distant metastases usually appear late in the course of the disease. However, in rare cases (as our case in discussion) and also especially in pediatric GISTs, distant metastases appears early and may present at diagnosis.[6]

CASE REPORT

We report on 29 years para 1 live 1 (P1L1), last childbirth 2 years (LCB), last menstrual period (LMP) 15 days, admitted to the institute of obstetrics and gynaecology (IOG) Egmore, Chennai with complaints of lower abdominal pain more on the right lower quadrant, vomiting and abdominal distension for 1 day. No history of constipation, hematemesis, melena and no history of any vaginal discharge. The patient had persistent tachycardia. Also, a history of the bilateral chocolate cyst was elicited and was on treatment for the same. No history of any previous abdominal surgery. No history of any medical comorbidities. Personal history-wise she had a good appetite, sleep and also her bowel and bladder were normal. She attained menarche at 15 years of age with a regular cycle thereafter and 2 years before she conceived spontaneously and gave birth to a female baby through full-term normal vaginal delivery.

She was admitted under a gynaecologist in IOG Chennai and was treated as a case of acute adnexal pathology. She was admitted to ICCU and was put on an acute abdomen protocol with a chart monitoring her pulse, BP, abdominal girth, SpO2, urine output, and periodic hemoglobin estimation. She was kept on nil per oral (NPO) and started on IV fluids, IV antibiotics, IV paracetamol and other supportive measures. After 12 hours of monitoring, as the pain didn't subside along with increasing tachycardia, a diagnosis of right torsion ovary based on USG (4×3 hyperdense in the right iliac region) finding was made and the patient was taken for emergency laparotomy by the gynaecology team.

The abdomen was opened by a pfannenstiel incision to repair the right torsion ovary. On opening the abdomen, the right ovary was found to be normal. Hence, the general surgery team was called in for per operative assistance. A thorough examination starting from the ileocaecal junction was made. The appendix was found to be normal. Approximately 70 cm from the ileocaecal junction a mass of about 15 cm was found. The mass is the intussusception of the ileum Figure 1. Both the intussuscepiens and intussusceptum were of ileal origin. As the intussusception region was congested, a decision to go for resection and anastomoses was made. The entire intussusception part was resected and an end-to-end four-layer anastomosis was done (Figure 2 and 3). After thorough irrigation, a pelvic drainage tube (DT) was kept and the abdomen was closed in layers.

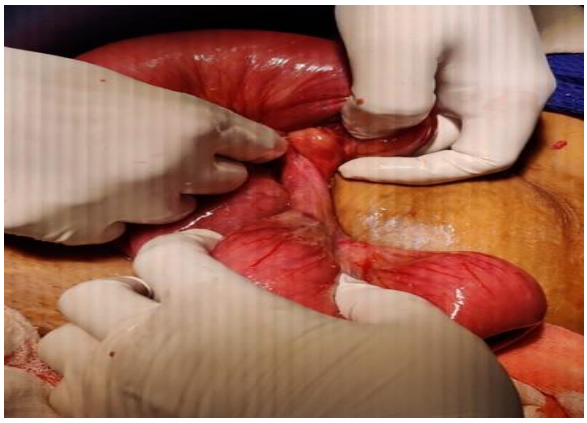

Figure 1: Intussusception on table.

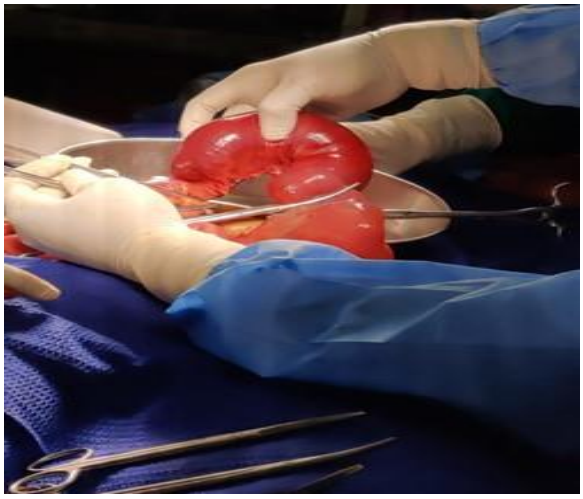

Figure 2: Resection of intussusception segment.

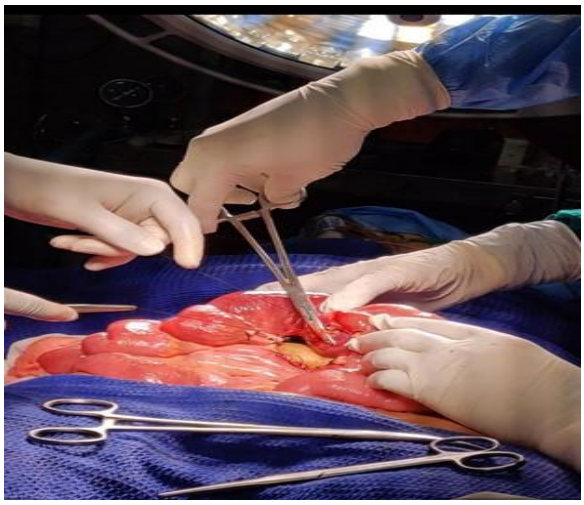

Figure 3: End to end anastomosis (ileum).

Post-op examination of the specimen revealed a nodule of size 2×2 cm in the intussusceptum, which appeared to the lead point in the evolution of the intussusceptum Figure 4 and 5. The total dissected bowel including both intussusceptions and intussusceptum was about 30 cm. The patient was subsequently taken over on the third postoperative day (POD) from the IOG, Egmore to the institute of general surgery, Madras medical college for further general surgical care. The post-operative period was uneventful. The patient was started on orals on the fourth POD, DT removed on the 6th POD and the patient discharged on the 8th POD with follow-up plans.

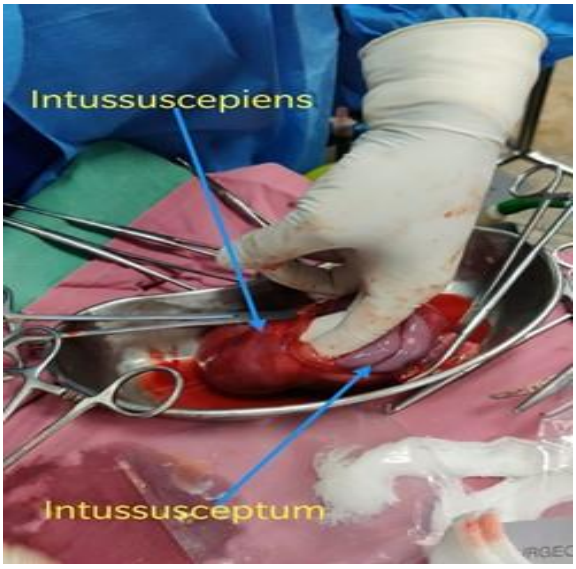

Figure 4: Dissected specimen 1.

The biopsy from the intussusception segment showed intestinal mucosa with ulceration and granulation tissue with an underlying neoplasm composed of spindle-shaped cells, a moderate amount of eosinophilic cytoplasm and elongated nuclei 1-2 mitoses/10HPF arranged in intervening fasciles and bundles admixed with scattered inflammatory cell infiltrate. The rest of the mucosa shows hemorrhage, ulceration and chronic inflammatory cells infiltrate and congested blood vessels. Both resected margins free of tumor. IHC CD117 was positive. An impression of CD117-positive GIST was given (Table 1).

Figure 5: Dissected specimen 2; GIST 2×2 cm presenting as lead point.

In the following metastatic work up a CECT abdomen (triple phase) was done. An ill-defined hypodense lesion of size 1.3×1 cm showing mild arterial phase enhancement in segment 4 (a) of the left lobe of the liver was observed. A similar lesion of size 1×1 cm in segment 7 of the right lobe of the liver identified and an impression of hepatic metastases in segments 4 (a) and 7 was given (Table 2). After proper counselling, the patient was sent to the medical oncology department to start her on imatinib, a tyrosine kinase receptor inhibitor which is the first line of the drug in the treatment of GISTs.

Table 1: Histopathology report.

Parameters	Inferences
Clinical diagnosis	Intussusception of ileum
Procedure done	Resection and anastomosis
Specimen	Resected bowel loop
Macroscopic appearance	Loop of bowel measuring 30 cm, nodule of size 2×2 cm present 5 cm away from one resected margin. The cut surface of the nodule appears smooth, glistening. The cut surface of the rest of the bowel appears normal.
Microscopic appearance	Intestinal mucosa with ulceration and granulation tissue with an underlying neoplasm composed of spindle-shaped cells the moderate amount of eosinophilic cytoplasm and elongated nuclei 1-2 mitoses/10HPF arranged in intervening fasciles and bundles admixed with scattered inflammatory cell infiltrate. The rest of the mucosa shows hemorrhage, ulceration and chronic inflammatory cell infiltrate and congested blood vessels. Both resected margins free of tumor.
IHC CD117	Positive
Impression	GIST arising from the small intestine causing intussusception.

Table 2: CECT abdomen triple phase.

Organs	Findings
Liver	Ill-defined hypodense lesion of size 1.3×1 cm showing mild arterial phase enhancement in segment 4 (a) of left lobe of liver; Similar lesion of size 1×1 cm in segment 7 of the right lobe of the liver.
Gall bladder	Normal
Spleen	Normal in size. Tiny, calcified granulomas
Pancreas	Normal
Adrenal glands	Normal
Kidneys	Both sides appear normal
Peritoneum	No evidence of free fluid in the peritoneal cavity. No evidence of peritoneal mass. No evidence of pre/para-aortic/para caval lymphadenopathy.
Bowel	Known case of ileoileal intussusception/post status ileoileal anastomosis. No evidence of wall thickening, No evidence of obstruction.
Aorta and IVC	Normal
Urinary bladder	Normal
Uterus	Copper T in situ appears normal.
Both ovaries	Normal
Impression	Known case of ileoileal intussusception/post status ileoileal anastomosis/biopsy proven GIST/ hepatic metastases in segment 4 (a) and 7.

DISCUSSION

In 2013 the WHO released an update of its 2002 classification system for tumors of the soft tissue and bone. The update incorporated more detailed cytogenetic and molecular data into the classifications. GISTs have been added in the update, with three subtypes benign, uncertain malignant potential and malignant.[7] Different grading systems are in vogue. The French federation of cancer centres sarcoma group (FNCLCC) system uses a three-grade system based on tumor differentiation, tumor necrosis and mitotic activity.[8] The national cancer institute (NCI) system also uses a three-grade system based on the evaluation of histology, location and tumor necrosis.[9] The American joint cancer committee/union for international cancer control (AJCC/UICC) grades GISTs separately from other sarcomas, using a two-grade system based on mitotic rate, as low grade (≤5 mitoses per 5 mm^2) and high grade (>5 mitoses per 5 mm^2).[10]

Both the European society for medical oncology (ESMO) and the national comprehensive cancer network (NCCN) follow the tumor-node-metastasis (TNM) classification of the AJCC/UICC. Based on the above guidelines the tumor in discussion comes under T1 N0 M1 which is stage IV. The NCCN recommends tyrosine kinase receptor inhibitor (TKI). The patient was subsequently referred to medical oncology and was started on imatinib (TKI). Lifelong continuation of TKA therapy is considered as an important component of continual care.

CONCLUSION

GISTs tumors less than 4 cm diameter are usually either asymptomatic or present with a nonspecific symptom. This case report shows, even a 2×2 cm GISTs can cause significant obstruction by evolving into an intussusception. This only adds to its protean presentation irrespective of the site and size of the tumor. GISTs tumor data on various presentations is limited. Continuous data collection including a multimodality type of approach involving early screening as in the Japanese health system will go a long way in the identification of GISTs in a very early stage. In addition to an early Screening protocol, doctors (general practitioners) and patient's education regarding the various types of presentations and symptoms, involvement of medical gastroenterology and medical oncology specialist in planning and managing complications of GISTs including early metastasis, will go a long way in the complete and comprehensive management of GISTs. Although various reports of 5 years survival after tumor-free resection for GISTs stands between 30-90% the median survival rate is 50-60% only. Patients treated with adjuvant therapy have a 46% lower risk of death than patients treated with surgery alone. Given the above observation, patient education regarding therapeutic intake of tyrosine kinase receptor inhibitors like imatinib and the need for lifelong clinical follow-up should be ensured to reduce mortality.

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